

Mozart's death: a rebuttal of Karhausen. Further evidence for Schönlein-Henoch syndrome

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This controversy, set in the late 19th century, is concerned not only with an evaluation of the eye-witness accounts of Mozart's relatives and friends, but also with the interpretation of the only two available medical documents. The question of the epidemic nature of the fatal illness is of crucial importance.

Mozart's swollen limbs

In her letter of 7 April 1825 to Nissen Sophie Haibel wrote: 'Now when Mozart fell ill, we both made him a night-jacket which he could put on frontways, since on account of his swollen condition he was unable to turn in bed.'^{1,2} Four years later, on 3 August 1829, Haibel informed Vincent and Mary Novello that Mozart's 'arms and limbs were much inflamed and swollen'³. Extracts of Sophie Haibel's letter to Nissen were published in his biography, while Otto Jahn obtained a copy of the letter from Ludwig von Köchel. Jahn expressed Sophie's narrative slightly differently: 'When he was taken ill we made him night-shirts which could be put on without giving him the pain of turning round'^{4,5}.

The term 'inflamed' is inappropriate for a painless peripheral oedema, which would not have caused difficulty for Mozart to turn over in bed. An acute polyarthritis is more in keeping with Joseph Eybler's description of the 'painful [schmerzvollen] final illness'⁶. Furthermore, Dr Guldener von Lobes' diagnosis of a rheumatic inflammatory fever [una febbre reumatico infiammatoria] leaves no doubt that there was an inflammatory affliction of Mozart's joints^{7,8}.

However Karhausen agrees that there was a progression of a generalized oedema which assumed the proportions of an anasarca⁹. Furthermore Karhausen concedes that if polyarthritis was present, it could be accounted for by the recurrent acute gouty arthritis, reported by Caner and Decker, in patients with chronic renal failure treated with periodic haemodialysis, hardly a predicament appropriate to this discussion!

An almost complete immobility

John Stone's presumption that my diagnosis of hemiparesis and stroke was based on a mistranslation in the English edition of Jahn in 1891, and the novelettish memoirs, attributed to Joseph Deiner, first published in Vienna *Morgen-Post* in 1856, is ridiculous¹⁰.

Nissen described Mozart's fatal illness: 'His final illness, in which he became confined to bed, lasted 15 days. It commenced with swelling of the hands and feet and an almost complete immobility [Einer beynahe gänzlichen Unbeweglichkeit] of them: after which followed sudden vomiting, which illness was

diagnosed as a heated miliary fever [Ein hitziges Frieselfieber]. Until two hours before he passed away he remained in full possession of his senses'¹¹.

It should be noted that Nissen listed Mozart's almost total inability to move as additional to, and separate from, the swelling of his hands and feet. The combination of polyarthritis and generalized oedema should not have caused an almost complete immobility. Edward Holmes translated it as 'an almost total incapacity of motion'¹².

Jahn expressed it slightly differently: 'Eine faft völlige Unbeweglichkeit', which Pauline Townsend translated as 'partial paralysis set in'^{4,5}. His friend Benedikt Schack stated that Mozart's weakness was such 'that he was obliged to be drawn forward whenever he required to sit up in bed'^{12,13}.

Admittedly the venesections would have aggravated Mozart's anaemia and uraemia, so adding to his weakness. Almost certainly Mozart would have been treated with laxatives to induce diarrhoea, which in association with vomiting would probably have caused hypokalaemia, which would have contributed to his weakness.

However I argue that for Mozart to have been unable to turn over in bed, or sit up without assistance, there must have been a muscular paralysis present. Further evidence for a paralysis is suggested by Joseph Eybler's account: 'I had the good fortune to enjoy his [Mozart's] constant friendship until his death; so that even in his painful final illness I helped to lift him up, lay him down, and wait upon him'^{6,14}.

There was no history of abrupt onset of weakness, which progressed rapidly over a period of days, to suggest a postinfectious polyneuritis such as Landry-Guillain-Barré syndrome, or a tick paralysis. Nor did the history suggest monoplegia or diplegia or paraplegia. I therefore concluded that the paralysis was most likely to have been a hemiparesis^{9,15}. Admittedly the combination of polyarthritis and anasarca per se might account for an almost complete immobility of Mozart's limbs.

The eye-witness account of Mozart's terminal coma was also recounted by Sophie Haibel to Nissen: '[Dr Closset] came and ordered cold poultices to be placed on Mozart's burning head, which, however, affected him to such an extent that he became unconscious and remained so until he died. His last movement was an attempt to express with his mouth the drum passages in the Requiem.'¹

Four years later Sophie informed the Novello's that she expressed her fears to Dr Closset 'that the sudden cold might be injurious to the sufferer, whose arms and limbs were much inflamed and swollen. But the doctor persisted in his orders and Madame Haibel accordingly applied a damp towel to his forehead.

Mozart immediately gave a slight shudder and in a very short time afterwards he expired in her arms.^{7,3}

It was stated in Nissen's biography: 'My sister-in-law thinks Mozart was not sufficiently well looked after in his illness, for instead of driving out the fever by other methods, they bled him and applied cold compresses to his head, whereupon his forces visibly forsook him and he lost consciousness, which he never again recovered.'^{7,11}

Jahn provided the following additional information: '[Closset] ordered cold bandages round the head, which caused such violent shuddering that delirium and unconsciousness came on, from which Mozart never recovered. Even in his latest fancies he was busy with the Requiem, blowing out his cheeks to imitate the trumpets and drums. Towards midnight he raised himself, opened his eyes wide, then lay down with his face to the wall, and seemed to fall asleep. At one o'clock (December 5) he expired.'^{5,16}

I agree with Dr Wheeler that the 'shuddering' which followed the application of the cold towels might have been shivering or rigors¹⁷. However I maintain that the sequence of events that followed is more in keeping with a convulsion following a stroke, such as a massive haemorrhage in either one of the frontal lobes or brain stem, causing facial nerve palsy and paralysis of conjugate gaze^{9,18}.

Un deposito alla testa

During their discussions of Mozart's case history Closset informed Dr Guldener von Lobes that he had diagnosed a 'deposito alla testa'^{7,8}. Closset had treated Constanze Mozart in July 1789, and had been attending Mozart since October 1791 or earlier. Closset had witnessed the evolution of the composer's neuropsychiatric symptoms of strange fainting fits, depression, and mental delusions. In diagnosing 'un deposito alla testa', literally a deposit in the head, Closset presumably suspected that Mozart was suffering from a serious intracranial lesion. However Closset was no doubt puzzled with the onset of a febrile illness associated with painful swollen joints, gross oedema, an almost complete immobility and a rash. On 28 November 1791 Closset called in consultation Dr Mathias von Sallaba, who diagnosed a heated miliary fever, the entry in the Register of Deaths^{9,15}. The recent research on the skull in the Mozarteum has raised the possible diagnosis of post-traumatic chronic extradural haematoma causing temporal lobe epilepsy as the cause of Mozart's strange fainting spells while composing *Die Zauberflöte* and the *Requiem*^{9,19}.

Epidemic Hitziges Frieselfieber

In his important testament, written on 10 June 1824, Dr Guldener von Lobes informed us: '[Mozart] fell sick in the late autumn of a rheumatic and inflammatory fever, which being fairly general among us at that time, attacked many people . . . This malady attacked at this time a great many of the inhabitants of Vienna, and for not a few of them it had the same fatal conclusion and the same symptoms as in the case of Mozart. The statutory examination of the corpse did not reveal anything at all unusual.'^{7,9}

It was recorded by Ignaz de Lucca (1746-1799) that there were 656 deaths in Vienna during the months of November and December in the year 1791, but details were not provided. In a separate entry the following deaths were recorded in 1791: stroke

(Schlag) 636, putrid fever (Faulfieber) 440, small pox (Pocken) 149, and accidental 64⁸. Many of the cases of putrid fever were presumably due to typhus²⁰.

Franz Niemetschek pointed out there was disagreement between Closset and Sallaba over the cause of Mozart's death²¹. Sallaba clearly recognized that Mozart's fatal illness was not a putrid fever. Furthermore, on clinical grounds, Mozart's retention of consciousness tends to exclude an enteric fever which some authors have proposed^{22,23}. Dr Karl von Bursy's diagnosis of 'Nervenfieber' probably referred to typhoid fever^{8,20,23}.

Karhausen's contention that there was no exanthem is inconsistent with Sallaba's diagnosis of 'Hitziges Frieselfieber'. Fortunately Professor Ferdinand Hebra, who was appointed to the Chair of Dermatology in Vienna in 1849, has written a detailed account.

The term 'Miliaria' ('Friesel' in German) referred to an eruption of distinct papules or vesicles, of the size of millet seeds - from whence the name was derived - or pins heads, or hemp seeds. Such descriptive terms as *Papulae miliaris*, or according to the colour of the vesicular fluid, *Miliaria rubra*, *M. alba*, and *M. crystallina* were in use²⁴.

In his treatise on *Der Friesel*, Franz Seitz outlined the diverse clinical spectrum which contrasted a sporadic exanthem like purpura (his term, not mine) at one end, extending through to a contagious disease, even occurring in endemic or epidemic forms, at the other. Such contagious cases were variously referred to under such titles as 'Miliaria substantiva exanthematica', 'Friesel-ausschlag', 'miliaria endemica', or 'Frieselfieber'²⁵. Hebra emphasized the absence of any subjective sensations of itching or tingling in the miliaria, so that the patient was often unaware of the eruption (was not this the case with Mozart?). Of the opinion that the miliaria were a manifestation of pyaemia, Hebra subdivided them into five categories: (1) typhus, (2) puerperal fever, (3) acute articular rheumatism, (4) the exanthemata, especially scarlet fever, and (5) a large miscellaneous group of inflammatory and febrile disorders²⁴.

There is then good evidence that in Mozart's Vienna it was recognized that certain contagious cases of 'Frieselfieber' were capable of erupting in endemic or epidemic outbreaks. Further evidence that Mozart's fatal illness was considered to be contagious was the fact that his distraught wife Constanze, after his death, lay down next to his corpse to catch his disease and die with him^{9,11}.

The validation of the epidemic nature of Mozart's fatal illness is of vital importance in this discussion since it not only dismisses the poisoning allegations but it also tends to exclude a diagnosis of infective endocarditis whose protean manifestations are otherwise compatible with Mozart's symptoms²⁶⁻²⁸.

The latter argument also tends to dismiss hyperthyroidism²⁹, obstructive uropathy - chronic pyelonephritis^{30,31}, and congenital renal tract anomaly^{17,32-34}. It is also of interest that Schönlein-Henoch syndrome has been noted as a complication of infective endocarditis³⁵.

Schönlein-Henoch syndrome: streptococci and epidemics

Karhausen has concluded erroneously that SHS is not a sequela of streptococcal infection and is not an epidemic disease. Beta haemolytic streptococcal infection was detected in one of two adult cases in

Gairdner's series³⁵, and in half of those tested in Cream's series of 77 cases³⁶. In other series mainly in children, the incidence of antecedent streptococcal infection has ranged from 21% to 33%³⁶⁻⁴⁰. In cases of Schönlein-Henoch nephritis an even higher incidence, ranging from 47% to 57% of those tested, was recorded^{41,42}.

In the two series where the incidence of preceding streptococcal infection in cases and controls was similar^{37,40}, the controls were not healthy asymptomatic children, but rather hospital inpatients matched by age and sex. The complex immunology remains ill understood today.

Five of Andrew's six cases were seen following an epidemic of scarlet fever⁴³. In the Danish epidemiological study, Nielsen concluded 'that Schönlein-Henoch purpura may be triggered by infection with several different micro-organisms, but there is no evidence that a single one such as the streptococcus is the major offender'. The incidence of a raised ASO titre among cases and controls was 28% and 16% respectively⁴⁴. In the Connecticut study it was concluded that a cluster of cases of SH purpura 'may have been caused by person-to-person spread of an infectious agent of the respiratory tract to susceptible hosts'. Fifty-five per cent of throat cultures from cases with a history of sore throat were positive for streptococci. Although the antibody evidence of recent streptococcal infection was similar in cases and controls, the authors concluded that 'it remains possible that recent streptococcal infection was one of the contributing factors in case children'⁴⁵.

Karhausen's preoccupation with the rarity of SHS in the 1990s is irrelevant to the scenario in Mozart's Vienna when streptococcal infections and their sequelae were a leading cause of morbidity and mortality. Furthermore, since the beginning of this century, there has been a decline not only in the incidence of such immune complex complications as rheumatic fever but also a decline in virulence of group A streptococci⁴⁶. Even as late as 1892 Osler described peliosis rheumatica (Schönlein's disease) as occurring most commonly in males between 20 and 30 years⁴⁷.

The cause of Mozart's death

Contracted during an epidemic, Mozart's fatal illness commenced on 20 November 1791 and lasted 15 days. The symptoms were pyrexia, polyarthritis, recurrent vomiting, exanthem, anasarca, near complete immobility and terminal coma¹⁵.

Most authors, other than the poisoning advocates and the rheumatic fever proponents, favour an underlying glomerulonephritis. While in the absence of an autopsy we will never be certain as to the cause of Mozart's death, the above features are all well accounted for only by the Schönlein-Henoch syndrome, which diagnosis is also favoured by other scholars^{48,49}.

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The Editor wishes to record thanks to all who have contributed to the debate on Mozart's illnesses during this centenary. A veil is now drawn across this particular stage. Future readers of the Journal will doubtless anticipate further enlightenment from future advances in medical knowledge at the next Mozart centenary AD 2056.